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Case Report

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A rare tumor cause of ileus: primary intestinal lymphoma

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ABSTRACT

Primary intestinal lymphomas (PBL) are rare tumors and extranodal lymphomas. They constitute 1% of malignant tumors of the gastrointestinal (GI) system. Colorectal lymphomas constitute 10-20% of all GI lymphomas and are most commonly located in the cecum. For a case to have PBL, in addition to the dominant GI symptoms, the clinical and radiological absence of pathological lymph nodes located outside the intestine, mediastinum, abdomen, or peripheral is required. In addition, there should be no findings in favor of lymphoma in the liver, spleen, and bone marrow. In this report, a 59-year-old female patient who was admitted to the emergency department with a clinical diagnosis of obstructive ileus while being investigated for abdominal pain lasting for 1 month, was operated on because of the development of an acute abdomen and an obstructive mass as a result of imaging, and was reported as having diffuse large cell lymphoma as a result of postoperative pathology, lastly as a result of the examinations performed, primary intestinal lymphoma was detected, and the diagnosis of the disease is presented. Since the first presentation of PBL may be intestinal obstruction and/or perforation due to delayed diagnosis, PBL should be kept in mind in patients presenting with obstructive ileus in emergency admissions.

Keywords: Primary gastrointestinal lymphoma, ileus, colorectal cancer

INTRODUCTION

Primary intestinal lymphomas (PBL) are extranodal lymphomas and are extremely rare tumors.[1] They constitute 1% of malignant tumors of the gastrointestinal (GI) system. Colorectal lymphomas constitute 10-20% of all GI lymphomas and are most commonly located in the cecum. Primary lymphoma of the colon, on the other hand, constitutes 0.2-1.2% of all colon malignancies.[2] The incidence is most common in patients over 50 years of age and is more common in women.[3] Abdominal pain (90%) is the most common clinical finding, and the others are abdominal mass (% 80), weight loss (45%), diarrhea, fever, loss of appetite, and constipation.[4] Obstruction is seen at a rate of 20-25% and perforation is less frequent.[5] We aimed to present our case of intestinal lymphoma due to its rarity. In this study, we aimed to present the case of intestinal lymphoma that admitted with GI tract symptoms clinic in contrast to classical lymphoma symptoms because of its rarity.

CASE

A 59-year-old female patient, who has no known additional disease other than diabetes mellitus, and has a history of surgery due to a previous knee and ovarian cyst, is being examined by the internal medicine clinic with outpatient follow-up due to abdominal pain that started 1 month ago. In the follow-ups, the patient has undergone a colonoscopy, and abdominal imaging is planned. During this period, the patient applied to our emergency department with complaints of sudden onset of abdominal pain, bloating, and inability to pass gas and stool.

In the physical examination; blood pressure 110/70 mmHg pulse: 90/min, Glasgow coma scale: 15, conscious, oriented, and cooperative. In the abdominal examination, there was a tenderness in the right lower quadrant, and no defense or rebound was detected. There was normal stool contamination on rectal touch.

In laboratory tests: White blood cell count (WBC): 13.2×10 (11.8 Neu), hemoglobin: 11.1 mg/dl, C-reactive protein (CRP): 30 mg/l, creatinine: 1.0 mg/dl, urea: 20 mg/dl, International normalized ratio (INR): 1.2.

In standing direct abdominal X-ray, there are 5–6 small intestine type air-fluid levels, in the contrast-enhanced abdominal tomography (CT) it was reported as "There is no evidence of intra-abdominal metastasis, asymmetric wall thickening at the level of the cecum, contamination in the surrounding adipose tissue, many millimetric lymph nodes, air-fluid levels in the ileal loops starting from the terminal

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ileum level, an appearance reaching a diameter of 34 mm, and there is also 4 cm of fluid in the pelvis." (**Figure 1**)

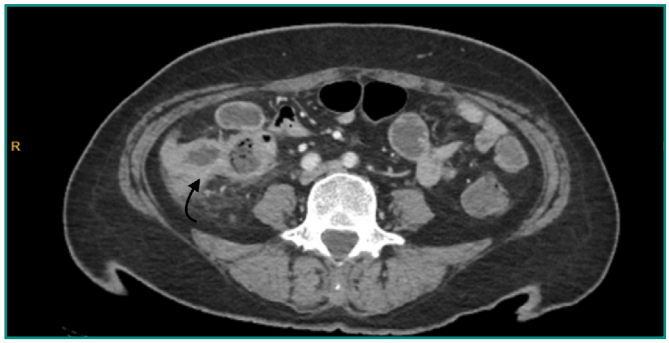

Figure 1. Abdominal tomography sections show irregular wall thickening (Black Arrow) at the level of the cecum and dilatation in the proximal small bowel loops.

Due to acute abdomen findings, severe abdominal distension and an occlusive lesion in the cecum, the patient was decided to have an emergency operation. During the exploration, no additional pathology was detected except for mass wall thickening at the level of the cecum and ileus, where the transition was at this level. The patient underwent right hemicolectomy and ileotransverse anastomosis (**Figure 2**). The patient, whose drain was terminated during the follow-ups, whose physical examination was normal, tolerated the regimen, and had discharged, was discharged on the 6th postoperative day.

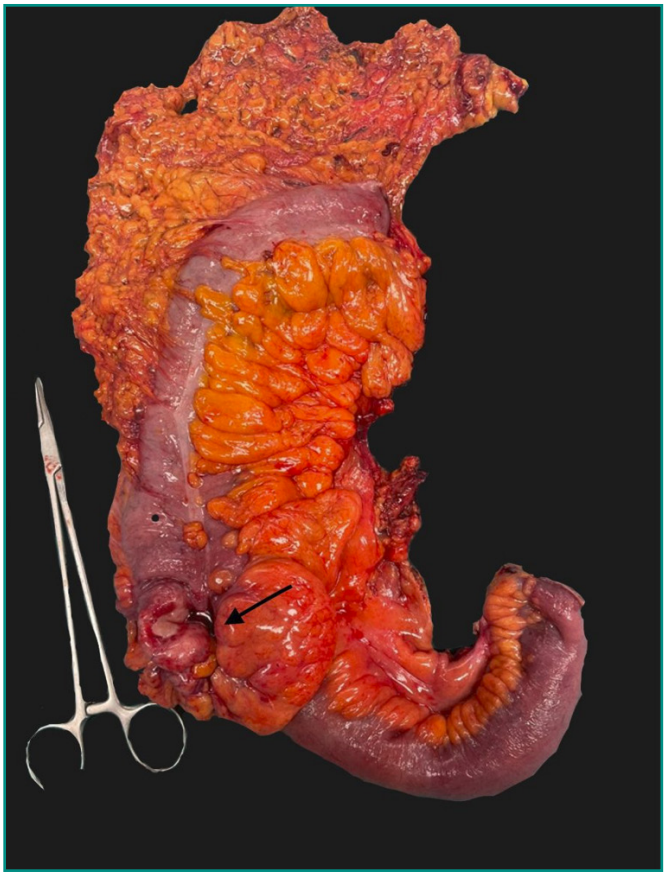

Figure 2. A right hemicolectomy surgical specimen and a tumoral mass located in the cecum (Black Arrow) are shown.

Pathology result; diffuse large cell lymphoma localized to the ileo-cecal region was reported. There was no involvement in the 35 resected lymph nodes and the appendix. In the immunohistochemical examinations, "90% with CD20 positivity ki67 40% with MUM1, 10-15% with bcl6; It was reported as "PankCK, CD3, CD10, bcl2 negative". No significant pathological uptake was detected in PET/CT taken for postoperative scanning, and the patient was transferred to the hematology clinic with the diagnosis of PBL. The patient was started on R-CHOP (rituximab, cyclophosphamide, adriamycin, vincristine, methyl prednisolone) chemotherapy by the hematology clinic.

Table 1. Diagnostic criteria for primary bowel lymphoma[11]
1- Absence of peripheral lymphadenopathy
2- Absence of pathologically mediastinal lymph nodes
3- Normal white blood cell count and peripheral smear
4- Normal bone marrow
5- The main lesion is in the gastrointestinal system and the pathological lymph node is in the area where the lesion drains
6- Absence of liver and spleen involvement

DISCUSSION

Its etiology is not known exactly, and it takes the 5th place among deaths due to malignancy.[6] In recent years, studies have found associations between intestinal lymphoma and immunosuppression after transplantation, celiac disease, inflammatory bowel disease, or human immunodeficiency virus (HIV) infection.[7] PBL is a heterogeneous group with different clinical and pathological manifestations, constituting the largest group with 4-12% of non-Hodgkin lymphomas and 40-50% of primary extranodal lymphomas.[8] PBL is located in the stomach (50-60%), small intestine (20-30%), colon, and rectum (10-20%), in order of frequency. Primary lymphoma of the colon constitutes 0.2-1.2% of all colon malignancies[9] and has a better prognosis than other malignancies (adeno CA, carcinoid, etc.).[5]

The most common symptom of GI lymphoma is abdominal pain; however, signs and symptoms are often nonspecific. In PBL, abdominal pain and vomiting are prominent in proximal cases, and abdominal distension is in distal localized cases. In the absence of disease-specific symptoms and intestinal obstruction, the diagnosis may be delayed.[1,2] Complications such as obstruction, perforation, bleeding, and intussusception may occur.[10] In our case, the patient admitted to the emergency room with non-specific gastrointestinal system symptoms in the last 1 month, and an emergency operation was decided without a definitive diagnosis due to obstructive symptoms.

Differentiation of primary lymphoma from secondary lymphoma is important because treatment approaches differ.[11] There are some criteria for accepting lymphomas seen in the GI system as primary. Peripheral smear and white blood cell were normal, peripheral lymph node and pathological mediastinal lymph node were absent, chest X-ray was normal, liver and spleen were not involved, and the lesion was limited to the gastrointestinal tract.

The cell type from which the tumor originates is generally B-cell (80-85%), as in the presented case, and to a lesser extent, T-cell (10-40%), which is reported to have a worse prognosis.[12] The prognostic features of the tumor; perforation, high grade histology, multiple mass and tumor stage.

It has been reported that the combined application of surgical resection of the mass and chemotherapy in PBL provides a successful remission of the disease and prevents complications.[13]

CONCLUSION

Unlike the classic lymphoma symptoms and clinic, PBLs can present unexpectedly. In conclusion, although rare, the diagnosis of PBL should be kept in mind for patients presenting with chronic GI system symptoms. Early diagnosis and complete resection without delay are important in terms of preventing morbidity and mortality.

ETHICAL DECLARATIONS

Informed Consent: All patients signed the free and informed consent form.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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